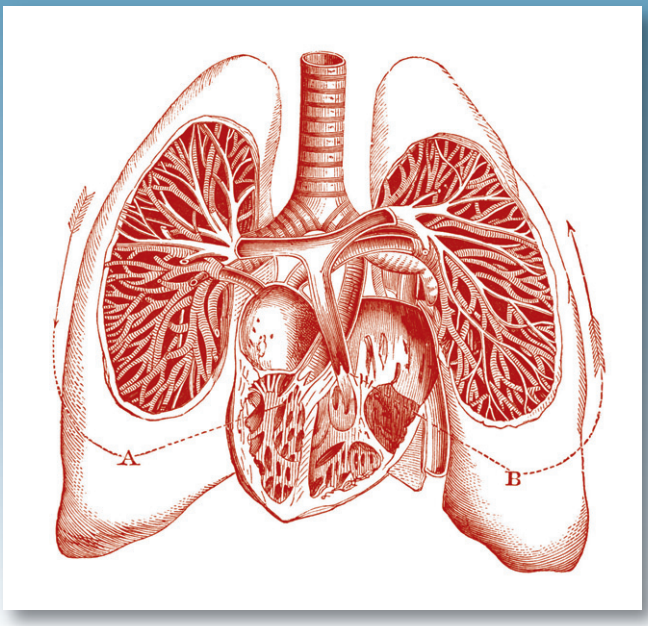

Evaluation and Management of the Adult Patient With Pulmonary Hypertension

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ABSTRACT

The prevalence of adult pulmonary hypertension (PH) in the United States has escalated in all races, gender, and ethnic populations. Nurse practitioners (NPs) are encountering adults with PH with increasing frequency. Through increased knowledge of the pathophysiological changes, diagnostic measures, and treatments for adults with PH, NPs collaborate with interdisciplinary teams to manage and improve client outcomes and quality of life.

Keywords: endothelin receptor antagonists, mean pulmonary artery pressure, phosphodiesterase inhibitors, prostanoids, pulmonary hypertension, right heart catheterization

Pulmonary hypertension (PH) is a progressive and ultimately fatal disease that presents as an elevated blood pressure in the pulmonary arteries. PH evolves as a comorbidity of other diseases or conditions, such as connective tissue diseases, lung diseases, liver disease, pregnancy, HIV infection, left heart failure, and the usage of the diet medication Fen-Phen, a combination of the 2 drugs fenfluramine (Pondimin) and phentermine (Lonamin). Although triggered by other diseases or conditions, the specific etiology of PH is not known. PH and comorbidity conditions affect a diverse segment of the population.

The demographic profile of adult PH patients includes men and women of all ages, racial, and ethnic groups. Between 2000 and 2002, the Centers for Disease Control and Prevention reported 807,000 adults were hospitalized with PH. Of these adults, 61% were women and 66% were 65 or older.[1] During the time period of 1980-2002, the number of PH deaths increased from 10,922 to 15,668, respectively; however, the greatest increase was observed only among women.[1] Among all racial populations, the number of adult PH deaths has increased mostly among the African American population.[1] Death rates are higher among the elderly, especially men 85 or older, women 65 or older, whites 75 or older, and African Americans 65 or older.[1]

Regardless of gender, age, or race, PH patients require continuous evaluation and management by an interdisciplinary team to alter disease progression and the resultant impact on multiple organ systems.

PH affects both the pulmonary and cardiovascular systems, with a potential negative impact on quality of life. PH patients have many acute and chronic health care needs that require meticulous evaluation, management, and care coordination. The nurse practitioner (NP) collaborates within an interdisciplinary team to effectively

evaluate and manage the PH patient and their comorbid health conditions.

PATHOPHYSIOLOGY

The pulmonary vasculature contains arteries and arterioles, which branch in the lungs to create a dense capillary bed to provide blood flow. The pulmonary capillary bed is a high-volume, low-pressure, low-resistance system that delivers blood to and from the lungs via the arterial and venous circulation systems, respectively. In a normal healthy adult, this system has a mean pulmonary artery pressure (MPAP) that ranges from 12 to 16 mmHg.[2]

Signs and symptoms of PH are related to the severity of right heart failure.

The dense capillary branching and the cumulative diameters of the capillaries provide a lower resistance pressure while effectively facilitating gas exchange.[3] The pulmonary vasculature system is effective during times of exercise as it can increase blood-carrying capacity by as much as 5 times with little or no increase in pulmonary pressure.[3] In a healthy individual the arterioles are thin-walled, distensible vessels that are able to dilate in times of increased metabolic need. The arterioles are composed of 3 layers: intima, media, and adventia.[4] The intima is the innermost layer and is responsible for tightening and relaxing the blood vessel, blood clotting, forming new vessels, inflammation, and immune responses.[4] The media is the middle layer that regulates the diameter and volume of the vessels, thereby distributing blood to areas of the body in need. The adventitia is the outermost layer and comprises collagen, which gives the vessel stability.[4] Chemical messengers, which include tumor necrosis factor and interleukin 1, serve as the mechanism of action that promotes the responsiveness of these arterial layers to increased metabolic needs.[2]

The pulmonary vasculature generates chemical messengers that produce, repair, and destroy cells in the vessel wall.[2] The blood vessels of the pulmonary circulatory system are controlled by a system of molecular messengers that facilitate the metabolic compensation through dilatation and constriction of these blood vessels. Prostacyclin, nitric oxide (NO), and endothelin (ET-1) are the molecular messengers that control the flow of blood in the pulmonary circulation. Prostacyclin is a prostaglandin that promotes blood vessel vasodilatation and prevents the overgrowth of cells in the blood vessels. Prostacyclin prevents platelets from clumping together, which lowers the incidence of clogged blood vessels. NO is a powerful vasodilator that prevents smooth muscle growth and inhibits platelets from sticking together. ET-1 is a messenger that causes vasoconstriction of the blood vessels. In healthy adults, these molecular messengers ensure appropriate compensatory blood flow based on metabolic demands.[4]

In adults with PH, smooth cells proliferate in the intima, thickening and clogging the arteriole walls. This altered tissue inside the arterioles forms a plexiform lesion. The lesion clogs the inside of the arterioles, causing scar formation of the intima and media arteriole layers. This narrows the vessel, with a resultant increase in MPAP. This scar tissue is fibrous and prevents the arterioles from expanding and contracting, thereby decreasing the normal compensatory mechanism of the pulmonary circulatory system. This thickening or remodeling of the intima arteriole layer is irreversible and leads to permanent vasoconstriction of the arterioles, with a resultant increase in MPAP.[4]

In addition, adult PH clients experience an imbalance among the molecular messengers contributing to pulmonary vascular remodeling, systemic hypoxia, and pulmonary thrombosis, which leads to increased MPAP and the development of PH.[2] The levels of the molecular messengers prostacyclin and NO, which produce vasodilatation throughout the pulmonary circulation and prevent platelet aggregation, are reduced, while concurrently there is an overabundance of the vasoconstricting properties of the molecular messenger ET-1. As a result of the imbalance in the molecular messengers, pulmonary vascular resistance (PVR) and MPAP are increased and negatively affect cardiac output.

A patient with PH has a resting MPAP $>$ 25 mmHg, with a pulmonary capillary wedge pressure (PCWP) $<$ 15 mmHg. As the disease progresses, right ventricular hypertrophy occurs as a result of the increased pulmonary vascular resistance (PVR), which leads to right heart failure, activity intolerance, and eventually can lead to death.[5] Signs and symptoms of PH are related to the